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DISEASES OF THE ESOPHAGUS

Review article

Dysphagia lusoria: a comprehensive review

B. Levitt, J. E. Richter

Division of Gastroenterology, Department of Medicine, Temple University School of Medicine, Philadelphia, PA, USA

SUMMARY. **Dysphagia lusoria is a rare vascular anomaly identified in a small number of patients being evaluated for dysphagia. The purpose of this paper is to present an illustrative case and provide a comprehensive review of the underlying anatomy, diagnosis, and management of dysphagia lusoria based on a review of the medical and surgical literature over the past 20 years.**

KEY WORDS: **dysphagia, lusoria, subclavian artery.**

Compression of the esophagus by a vascular structure is an uncommon cause of dysphagia. There have been reports of almost every major vasculature structure within the chest cavity causing some degree of esophageal compression and symptomatic dysphagia.[1–3] In 1794, David Bayford noted the association of dysphagia with esophageal compression in a 62-year-old woman caused by a right aberrant subclavian artery. He coined the term 'dysphagia lusus naturae', which is Latin for 'freak of nature.' An aberrant right subclavian artery is the most common congenital anomaly of the aortic arch, with an incidence ranging from 0.5% to 1.8%.[4,5] We present an interesting case of dysphagia lusoria, and based on a Medline and Pubmed review of the English medical and surgical literature over the past 20 years will discuss the anatomy, diagnosis, and management of this uncommon disorder in adults.

CASE

The patient was a 24-year-old healthy woman referred to the Esophagus Clinic at our hospital (Temple University Hospital, Philadelphia, PA, US) for a second opinion of her chronic swallowing problems. She described lifetime symptoms of postprandial chest pressure and spasm, with a sensation of swallowed air sticking in her mid-upper chest within minutes after eating. These symptoms worsened with carbonated beverages, but otherwise were not related to any other specific foods. Her symptoms improved when she lay supine. There was no dysphagia for foods, but occasionally large multivitamin pills stuck in her mid-upper chest. No odynophagia, nausea, vomiting, regurgitation, or weight loss were present, but the patient noted intermittent heartburn that improved with proton pump inhibitors. No other medications, including aspirin or non-steroidal anti-inflammatory drugs, were used by the patient.

Her physical examination, including palpation of her peripheral pulses, was unremarkable. By report from her primary gastroenterologist, the patient had a negative upper endoscopy and barium esophagram. Unfortunately, the esophagram was not available for our review. A barium esophagram performed at Temple University Hospital demonstrated an oblique ascending extrinsic compression above the level of the aortic arch (Fig. 1). A 13 mm tablet hung-up at the region of the external compression, replicating many of the patient's symptoms. CT scan imaging demonstrated an aberrant right subclavian artery compressing the esophagus as it traversed posteriorly (Fig. 2). There was no evidence of a Kommerell's diverticulum or aneurysmal dilation.

A lengthy discussion ensued in which the different management strategies were outlined. Given the atypical nature of her symptoms, as well as her stable weight, consideration was given to providing symptomatic relief with lifestyle changes, together with a

Address correspondence to: Joel Richter, MD, Temple University School of Medicine, 3401 N. Broad Street, 807 Parkinson Pavilion, Philadelphia, PA 19140.
Email: jrichter@temple.edu

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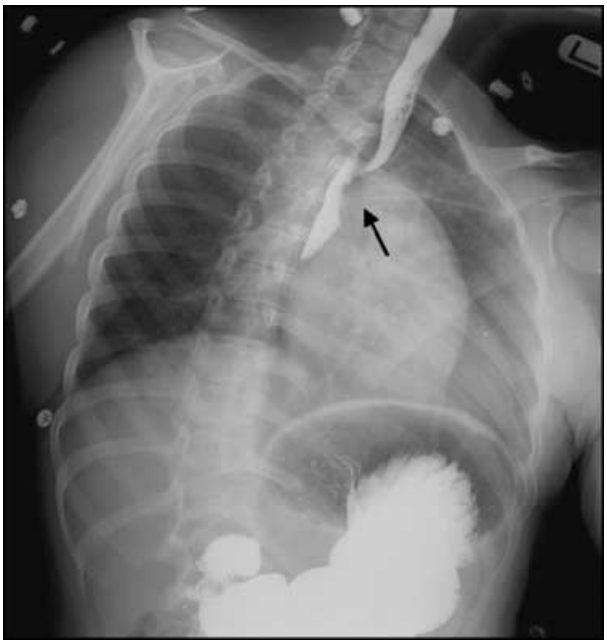

Fig. 1 Barium esophagram demonstrating an oblique ascending extrinsic compression above the level of the aortic arch.

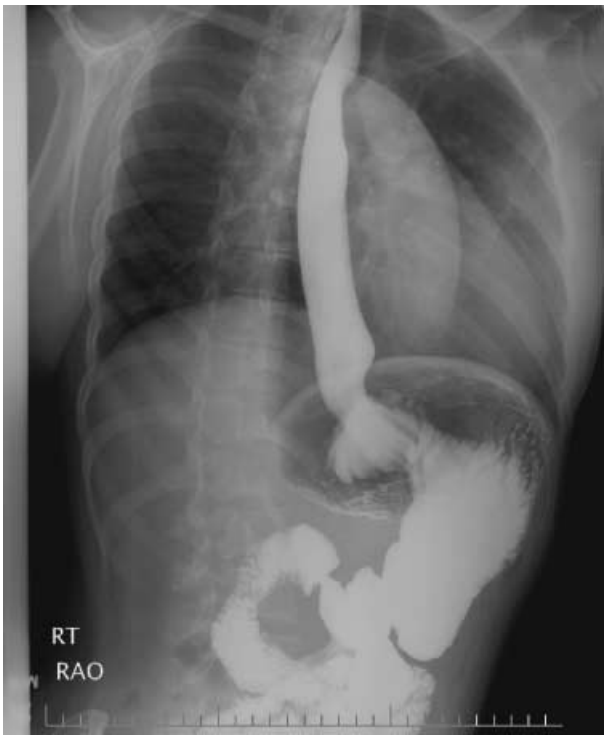

Fig. 3 Follow-up barium esophagram demonstrating normal postoperative esophageal contour.

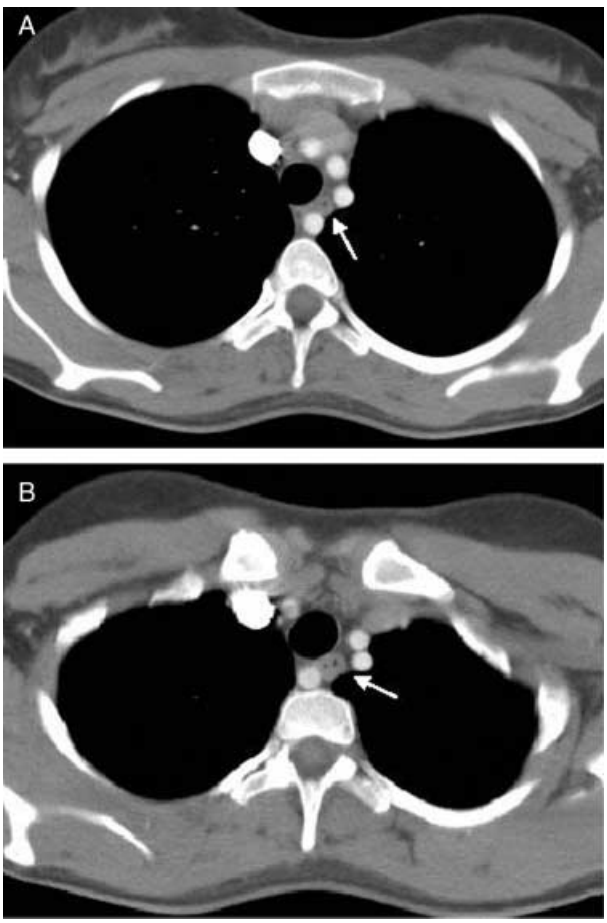

Fig. 2 CT scan of the thorax demonstrating: (A) posterior esophageal compression by aberrant vessel; and (B) re-expansion of the esophagus distal to site of compression.

possible trial of medications, such as proton pump inhibitors. The patient felt her symptoms were greatly impacting her quality of life, and therefore wanted to discuss her surgical options. She was referred to vascular surgery. After discussing the surgical procedure and its potential complications, the patient decided to proceed with surgery. A pre-operative angiogram was consistent with the prior CT scan findings, demonstrating an aberrant right subclavian artery. The surgeons stabilized the aberrant artery via a supraclavicular approach, after which a transposition of the subclavian to the common carotid artery was performed. The aberrant artery was ligated at its aortic arch takeoff through a limited thoracotomy.

Upon follow-up 3 months after the surgery, the patient was about 50% better. Pills no longer stuck in her chest, and the sensation of spasm was improved. However, the postprandial chest pressure persisted. A follow-up barium esophagram was normal, with resolution of the extrinsic compression (Fig. 3), and a 13 mm tablet passed rapidly into the stomach. An endoscopy and esophageal manometry were unremarkable.

PATHOGENESIS

Developmental anomalies of the aortic arch and its major branches are relatively common, being found in 3% of people in autopsy studies. The 'jest of nature'

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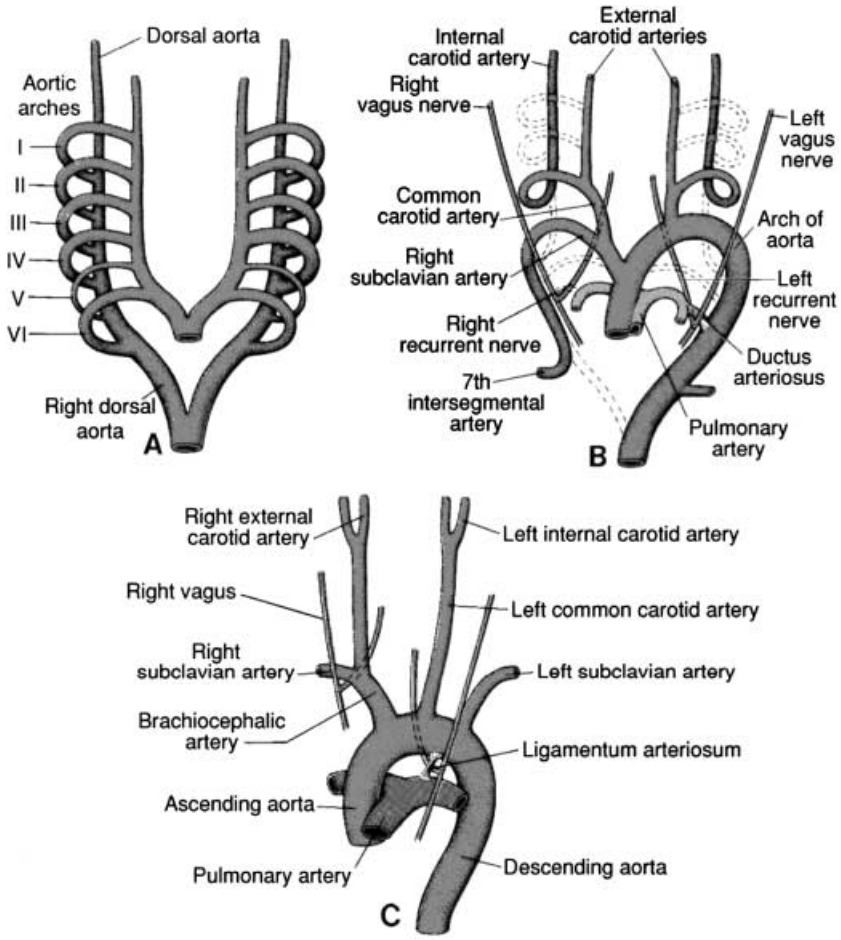

Fig. 4 Typical development of the great vessels. (A) In early in-utero development, the aortic arches are a bilateral duplicate system. (B) As the great vessels develop, the right aortic arch system undergoes atresia. (C) Fully developed anatomy of the great vessels at birth.

described by Bayford is a birth defect encompassing any aortic root vascular anomaly that causes esophageal dysphagia. During normal embryologic development, the aortic arch begins as a duplicate system. The right aortic arch becomes atretic proximal to the take-off of the right subclavian artery and the right common carotid artery. These vessels normally merge to form the right innominate artery, which is typically the first branch off the left aortic arch. The three vessels classically emerging from the aortic arch in the developed fetus are the innominate artery, which branches into the right subclavian and common carotid arteries, the left common carotid artery, and the left subclavian artery (Fig. 4).

The specific embryologic abnormality of the aortic arch responsible for an aberrant right subclavian artery is involution of the fourth vascular arch, along with the right dorsal aorta, leaving the seventh intersegmental artery attached to the descending aorta (Fig. 5). Since the persisting right aortic arch forms the root of the aberrant artery, the artery often has a broad base, referred to as a 'Kommerell's diverticulum.'[6] This persistent intersegmental artery assumes a retroesophageal position as it proceeds out of the thorax into the right arm. In 80% of cases, it crosses between the esophagus and the vertebral column, in 15% of cases it runs between the esophagus and the trachea, and in 5% of cases it passes anterior to both the trachea and esophagus. Some reports suggest this anomaly only produces symptoms in the setting of a concomitant congenital anomaly of the carotid arteries, either a common trunk to both carotid arteries ('bicarotid truncus,') or a close origin of the carotid arteries, leading to decreased distensibility of the esophagus as it is compressed posteriorly by the aberrant vessel.[7] However, this specific abnormality is absent in many reported cases of dysphagia lusoria.

There is a higher prevalence of vascular abnormalities, up to 37%, and specifically an aberrant right subclavian artery, in children with Down's syndrome and congenital heart disease. The abnormality may be seen in up to 2% of patients with tetrology of Fallot, pulmonary atresia, and major aortico-pulmonary collateral arteries.[8] Other vascular anomalies causing dysphagia include a persistent right aortic arch with aberrant left subclavian artery, a tortuous or aneurysmal thoracic aorta (dysphagia aortica), and left atrial enlargement.[1–3]

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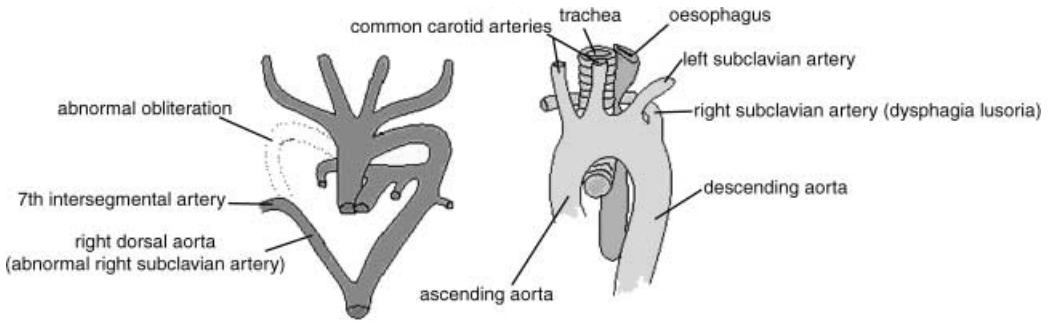

Fig. 5 Embryologic defect leading to aberrant right subclavian artery. Please note the position of the esophagus between the trachea and aberrant right subclavian artery.

CLINICAL PRESENTATION

The presence of this anomaly is often asymptomatic, and may be discovered incidentally on imaging or on postmortem analysis. As many as 60–80% of patients remain symptom-free in their lifetime. Symptomatic adult patients usually present with dysphagia consistent with a mechanical obstruction. Symptoms are primarily for solids and are associated with regurgitation of unchewed food, postprandial bloating, chest pain, and symptoms that frequently change with position. Other complaints include coughing, thoracic pain, or Horner's syndrome. In rare cases, patients present with rupture of an aneurysmal aberrant artery or Kommerell's diverticulum.[9] In our literature review, we found that the presenting symptom in 91% (31/34) of reported cases was dysphagia. Less then 20% of the patients complained of chest pain. Only one patient presented with an aneurysmal rupture.

In infants, respiratory symptoms are the predominant mode of presentation. This is believed due to the absence of tracheal rigidity, allowing for its compression and leading to stridor, wheezing, cyanosis, or recurrent pneumonia. Interestingly, patients usually do not present in childhood, becoming symptomatic in young adulthood, and even in the middle or elder ages. The average age of patients in our review was 48 years. Theories explaining this delayed presentation include physiologic and anatomic changes that may occur with the aging process such as increased esophageal rigidity, rigidity of the vessel wall due to atherosclerosis, elongation of the aorta, and aortic aneurysm formation, especially in the presence of a Kommerell's diverticulum.[10–12]

DIAGNOSIS

The physical examination is usually normal. Asymmetrical radial pulses may be present, especially in the setting of other vascular abnormalities.

Upper endoscopy is usually normal. Occasionally, a pulsating extrinsic compression of the posterior wall of the esophagus may be present. In our review,

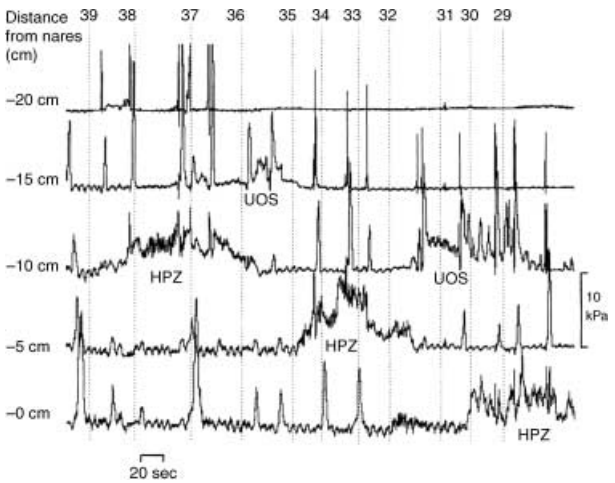

Fig. 6 Esophageal manometry showing high pressure zone at 26–29 cm, 10 kPa = 75 mmHg.

five patients were described as having this abnormal finding on upper endoscopy. However, many papers do not report the performance of an upper endoscopy, so the true incidence of this abnormality cannot be accurately derived. Some authors have described the loss of the right upper extremity radial pulse with compression of the pulsating mass by endoscopic manipulation, as well as enhancement of the midesophageal luminal compression with arm elevation.[1] Endoscopic ultrasound, especially with the use of Doppler technology, can be useful to verify the vascular nature of an abnormality seen on endoscopy.

Esophageal manometry frequently reveals nonspecific findings. Some report a high-pressure zone at the site of impingement by the lusorian vessel, with superimposed pulsation synchronous with the arterial pulse. Additionally, high peristaltic pressures in the proximal esophagus above the level of the compression may be present (Fig. 6).[1,3,10,13] Our review of the literature found that 5/34 patients had this manometric abnormality. However, once again, many reports do not detail esophageal manometry results, making the true incidence of this finding unknown. Most authors investigating the role of

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esophageal manometry in evaluating patients with dysphagia lusoria have concluded that there are no pathognomonic manometric findings that are useful in diagnosing the syndrome or predicting the need for surgery.[1–3,10]

The best method to diagnose an aberrant right subclavian artery presenting with dysphagia is initially with a barium esophagram followed by either a CT or MRI scan. Barium esophagram typically shows an oblique ascending extrinsic compression above the level of the aortic arch (Fig. 1). It may be helpful to administer a tablet or other solid bolus to improve localization of the defect, and occasionally reproduce the patient's symptoms. In our literature review, 14/34 patients had an abnormal barium esophagram. In the remaining 20 cases, there was no report of the performance of a barium swallow. This omission likely reflects the fact that the study was not performed, rather than the patients having had normal barium X-rays. The diagnosis can easily be missed when the high thoracic esophagus is not carefully examined, and when lateral or oblique views of the esophagus are not obtained.

In the past, angiography was the gold standard to confirm the diagnosis of vascular anomalies. With today's technology, CT angiography or MR angiography have largely supplanted the role of conventional angiography. Both of these modalities are capable of clarifying the anatomy of the aortic arch, and its relationship to surrounding organs. Additionally, these imaging techniques are helpful in diagnosing any other intrathoracic pathology that may be present. With the advent of volume rendering, and 3-D reconstructions, the anomalies of the aortic anatomy can be even better visualized. In a recent study evaluating the reliability of multidetector CT scan to detect an aberrant subclavian artery, Alper *et al.* found vascular pathology in 15/38 patients (40%) presenting with dysphagia who did not have abnormalities on any other investigative study.[14]

MANAGEMENT

The management of patients with dysphagia lusoria primarily depends on the severity of symptoms. Mild to moderate symptoms are often treated symptomatically, with changes in lifestyle and dietary modification, such as avoiding exacerbating foods, eating slower, chewing well, taking smaller bites, sipping liquids, as well as reassurance. Additionally, acid suppression and promotility agents have been used. In a series of six patients, Janssen *et al.*[10] were able to treat three with a proton pump inhibitor alone, or in combination with the prokinetic drug cisapride. Whether or not this indicates that the dysphagia associated with this vascular anomaly is secondary to underlying gastroesophageal reflux disease or a motility disorder, and the vascular anomaly is merely an incidental finding, is a matter of speculation.

Other patients do not respond to conservative therapy, and in these situations, surgical intervention is justified. In Janssen's paper, three patients required surgical intervention. In our literature review, an additional 14/24 patients underwent surgical repair of the aberrant vessel. In the remaining seven cases, the treatment course was not provided. Additionally, many of the cases were in the surgical literature, so it is not clear whether the patients undergoing surgery failed medical therapy prior to surgical intervention.

The surgical approach largely depends on the specific vascular anatomy as well as the surgeon's personal preference. The goal of surgery is to remove the aberrant vessel and to reconstruct the vessel in its appropriate position. The reconstruction can be done via anastomosis of the native vessel, or by interposing a synthetic graft in its place. There does not appear to be a general consensus in the surgical literature as to the best surgical approach. The traditional approach consists of dividing the aberrant artery at its origin through a median sternotomy and translocating the distal subclavian artery to the aortic arch or right carotid artery. A cervical approach is preferred because there is a decreased rate of complications, as well as better visibility of the subclavian and carotid arteries. Through a cervical approach, the aberrant artery is ligated near its root and connected with the right carotid artery. There are even reports of severing the artery without reconstruction, without significant reports of upper extremity ischemia. Potential damage to nearby structures and/or altering blood flow to the right upper extremity are possible complications. In the past, mortality rates have been reported to be as high as 16–25%, largely due to a two-step surgical repair.[15] With newer surgical techniques, more recent reports quote mortality rates as low as 0%.[16]

In patients who are poor surgical candidates, endoscopic dilation of the esophageal narrowing may temporarily relieve symptoms of dysphagia.[17] There are not enough cases in the literature to know the success rate of this procedure. However, its utility is primarily palliative in patients who cannot undergo more definitive therapy. Alternatively, there are various procedures that interventional radiology can perform in an effort to endovascularly ligate the aberrant vessel while maintaining blood flow to the appropriate organs.[18]

SUMMARY

In summary, dysphagia lusoria caused by an aberrant subclavian artery is a rare cause of dysphagia in adults. The ideal method of diagnosis includes

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barium esophagram with confirmatory CT angiography or MR angiography. The ideal management strategy is unclear, but it seems that in patients with mild to moderate symptoms, lifestyle changes and dietary modification, with possible medical therapy, is indicated. In more severe cases, surgical intervention should be considered. Our case is an example of a patient with vague, atypical symptoms who opted for surgical intervention with mixed results.

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